

Case Report :
LHERMITTE'S SIGN AS THE PRESENTING SYMPTOM OF
VITAMIN B₁₂ DEFICIENCY

by

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Vitamin B₁₂ deficiency is a potential cause of permanent neurological damage. Early diagnosis and treatment with hydroxocobalamin may reverse the clinical signs and prevent permanent neurological damage.

A 47-year-old garage proprietor presented with a three-month history of numbness and tingling of his feet and subsequently of both hands. These symptoms were accompanied by a sensation of electric shocks in his arms and legs, precipitated by forward flexion of the neck. There was no history of trauma to the cervical spine and he had previously been in good health.

General physical examination was normal. There was no abnormality of the cranial nerves and no limitation of neck movement. No weakness or wasting of the limbs was present. There was no impairment of light touch, pin-prick, joint position, temperature or vibration sense. The plantar responses were flexor, with normal deep tendon reflexes and gait. Investigations revealed haemoglobin 13.6 g/dl, a mean corpuscular volume of 105 fl (normal 80-96) and a white blood cell count of $4.9 \times 10^9/l$. The serum folate was 27 nmol/l (normal 4.5-34.0) and the serum vitamin B₁₂ by radioisotope assay on three separate occasions 162, 133 and 213 pmol/l (normal 88-664). Bone marrow examination showed the presence of megaloblastic erythropoiesis. X-ray of the cervical spine in flexion showed moderate anterior subluxation of C₄ on C₅ and to a lesser extent C₅ on C₆. A metrizamide myelogram demonstrated only minor disc protrusions at C_{3/4} and C_{5/6}. Cerebrospinal fluid cells and protein were normal. The Schilling test confirmed a B₁₂ absorption defect which was corrected with intrinsic factor. The serum gastrin was 1050 pg/ml (normal 0-100) in keeping with achlorhydria. Antibodies to gastric parietal cells and intrinsic factor were present. Following treatment with injections of hydroxocobalamin 1 mg daily for seven days and monthly thereafter, the patient's Lhermitte's sign and sensory symptoms resolved completely over the subsequent four months. At follow-up one year later he remains well with a normal mean corpuscular volume of 90 fl.

DISCUSSION

Lhermitte's sign, a sensation of sudden tingling or electric discharge down the spine and limbs on flexion of the neck, was first described following trauma to the head by Marie and Chatelin in 1917,¹ and after trauma to the neck by Babinski and Dubois in 1918.² In 1924 Lhermitte et al³ described a patient with disseminated sclerosis presenting with this symptom, but it was not until 1933 that its occurrence in sub-acute combined degeneration of the spinal cord was reported.^{4, 5}

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It is generally accepted that Lhermitte's sign results from damage to the sensory pathways in the cervical cord and is most commonly found in multiple sclerosis and also in vitamin B₁₂ deficiency.^{6, 7} The common pathological mechanism of these two conditions is demyelination which has been well described in vitamin B₁₂ deficiency.⁸ Animal studies confirm that demyelinated axons are stimulated by mechanical deformation.⁹ In the human, similar deformation of the cervical cord occurs during neck flexion resulting in excitation of demyelinated axons.

The present case is remarkable in that Lhermitte's sign was encountered without any indications of posterior column disease and with a normal vitamin B₁₂ level. We postulate that the moderate co-existent subluxation of the cervical spine during flexion deformed the spinal cord more than usual, and thus generated excitation in only mildly demyelinated sensory neurones. Treatment with vitamin B₁₂ reversed the demyelination, resulting in clinical resolution of the dysaesthesia and Lhermitte's sign, despite the persistent bony abnormalities of the cervical spine.

It has been recognised for many years that the neurological complications of vitamin B₁₂ deficiency may precede or occur without haematological abnormalities, but it is also becoming apparent that neurological disease may occur with normal vitamin B₁₂ levels.¹⁰ This case confirms that a normal vitamin B₁₂ level does not exclude vitamin B₁₂ deficiency, and demonstrates the importance of proceeding to further investigation including a Schilling test if early diagnosis and treatment are to prevent irreversible neurological damage. Patients presenting with Lhermitte's sign who do not have clear evidence of multiple sclerosis should have vitamin B₁₂ deficiency excluded.

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